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DRESS syndrome

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DESCRIPTION

A 16-year-old boy, a known case of epilepsy on oral phenytoin (300 mg/day) and folic acid (5 mg/day) for last 6 weeks, presented with complains of high-grade fever, jaundice and generalised erythematous eruption for 15 days duration. The eruption was maculopapular at onset, which desquamated in the next 3 days and changed to exfoliative dermatitis (figure 1). The patient was on no other medication. He had generalised tender lymphadenopathy, jaundice and tender hepatomegaly. Investigations revealed haemoglobin 12 g/dl, total leucocyte count 15 700/mm³, differential leucocyte count revealed eosinophils 15%, polymorphs 52%, lymphocytes 28% and monocytes 5%. Serum bilirubin was 7.6 mg/dl (0.3–1.0 mg/dl), alanine aminotransferase and aspartate aminotransferase were more than five times the normal. Biopsy of a skin rash revealed spongiosis with intraepidermal vesiculation and patchy exocytosis with no specific pathology in the dermis. All other investigations including septicaemia profile and serology for viral hepatitis were negative. Diagnosis of drug rash with eosinophilia and systemic symptoms (DRESS) syndrome due to phenytoin was made. Phenytoin was discontinued, and patient was started on oral prednisolone at the dose of 1 mg/kg/day for 2 weeks, which tapered over a period

of next 4 weeks. The patient showed rapid resolution of fever, eosinophilia and progressive improvement in skin rash and liver dysfunction over a period of 3 weeks. Our patient was diagnosed as DRESS syndrome as defined by Bocquet *et al.*¹ Clinical features were typical: fever, rash followed by exfoliative dermatitis, lymphadenopathy, eosinophilia and hepatitis. Drugs that commonly cause DRESS syndrome include phenytoin,² phenobarbital, carbamazepine, lamotrigine, minocycline, sulphonamides, sulphasalazine, trimethoprim, allopurinol, abacavir, nevirapine, mexiletine, isoniazid, gold salts, diltiazem, atenolol, captopril, azathioprine and dapsone. DRESS syndrome usually begins several weeks after exposure to the offending drug. The overall mortality in DRESS syndrome is about 10%.¹ The most common differential diagnoses for DRESS syndrome are Stevens-Johnson syndrome/toxic epidermal necrolysis, hypereosinophilic syndrome and Kawasaki disease.

Learning points

- ▶ Drug rash with eosinophilia and systemic symptoms (DRESS) syndrome is a life-threatening adverse effect of aromatic anticonvulsants (phenytoin, phenobarbital and carbamazepine).
- ▶ Clinicians should have a high index of suspicion for the DRESS syndrome in patients being treated with aromatic anticonvulsants who present with sepsis-like syndrome.
- ▶ Management of DRESS syndrome is recognising the presence of this syndrome, immediately stopping the offending drug and use of steroids.

Competing interests None.

Patient consent Obtained.

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Figure 1 Exfoliative dermatitis over both lower limbs.

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